CASE REPORT

Hiding in plain sight; constrictive pericarditis

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SUMMARY

We present a case of constrictive pericarditis that was especially difficult to diagnose. The patient presented with generalised oedema, dyspnoea and pleural effusions. History was significant for prior polysubstance abuse but was otherwise unremarkable. Physical examination revealed only jugular venous distention. CT demonstrated a normal pericardium with pleural effusions. Echocardiography showed mildly elevated right ventricular pressures with dyssynergic motion of the ventricular septum. No intervention was being carried out, but 1 month later further evaluation with a right and left heart catheterisation showed the classical square-root sign with equalisation of diastolic pressures in both ventricles as well as ventricular interdependence. Idiopathic constrictive pericarditis was thus diagnosed with a subsequent pericardial stripping which confirmed a thickened pericardium encasing the heart.

BACKGROUND

This patient did not have any risk factors such as thoracic radiation, chemotherapy, thoracic surgery or tuberculosis. Imaging with both echocardiography and CT did not reveal a thickened pericardium. It was not until a high index of suspicion and subsequent cardiac catheterisation that a diagnosis was made. This case highlights the need to consider an early diagnosis of constrictive pericarditis, in spite of no risk factors, as most cases are idiopathic in nature.

CASE PRESENTATION

A 48-year-old man with no significant medical history presented with a 3-week history of acute onset of dyspnoea on exertion progressing to dyspnoea at rest and associated with paroxysmal nocturnal dyspnoea, orthopnoea and peripheral oedema. He denied fever, cough or chest pain. Vital signs were stable with an oxygen saturation of 98% on room air. Physical examination demonstrated jugular venous distension and dullness to percussion bilaterally at the lung bases. A chest x-ray revealed bilateral pleural effusions. ECG showed normal sinus rhythm with non-specific T-wave abnormalities. Echocardiogram was significant for dyssynergic movement of the septum and mildly increased right ventricular systolic pressures (RVSP) at 33 mm Hg. Ejection fraction was preserved, no valvular abnormalities were seen and the pericardium was unremarkable. Thoracentesis revealed an exudative effusion. He was subsequently diagnosed with pneumonia and treated with levofloxacin and diuretics. There was an initial improvement in the symptoms; However, the patient required recurrent thoracentesis several months later. Exudative effusions with predominant lymphocytes and negative cytology were

noted. Further investigations showed negative Gram stain and negative cultures for bacteria, fungus and acid-fast bacilli. Tuberculin-purified protein derivative test was also negative. Repeat echocardiogram was unchanged. Six months after the initial presentation the patient presented complaining of severe dyspnoea at rest associated with generalised oedema. Physical examination now showed normal vitals, jugular venous distention with no Kussmaul's sign, distant heart sounds with no murmurs or rubs and decreased breath sounds in the inferior 2/3rd of the right lung with dullness to percussion. There were no signs of ascites or hepatosplenomegaly. Pitting oedema was present in both the lower extremities extending to the sacrum and the abdominal wall.

INVESTIGATIONS

Repeat CT showed stable appearance of the thorax with a moderate-to-large right pleural effusion, right lower lobe atelectatic changes with superimposed consolidation versus airspace disease and a small effusion on the left. A small amount of perihepatic ascites was present and was slightly increased since previous examination. No calcification or effusion within the pericardium were noted. Complete blood count (CBC) was unremarkable, comprehensive metabolic panel revealed borderline low Na at 135 mmol/l (135–145 mmol/l), elevated total bilirubin 1.6 mg/dl (0.1–1.1 mg/dl) with elevated direct bilirubin 0.6 mg/dl (0.0–0.3 mg/dl), elevated γ -glutamyl transferase at 276 units/l and elevated alkaline phosphatase at 283 units/l (20–130 units/l). Cardiac enzymes were normal and brain natriuretic peptide (BNP) was slightly above normal at 175 pg/ml (0–125 pg/ml). Urine analysis showed elevated protein 22.3 mg/dl (0.0–11.8 mg/dl) and elevated specific gravity 1.023 (1.008 to 1.022). Duplex ultrasound of the lower extremities was negative for deep venous thrombosis. Owing to recurrent pleural effusions of unknown origin a right-sided video-assisted thoracoscopy (VATS) with pleural biopsy and talc pleurodesis were performed which removed 2700 cc of yellow pleural fluid. Frozen section performed during the time of surgery revealed chronic lymphocytic pleuritis with no malignancy. Repeat echocardiogram showed an ejection fraction of 55–60% with no regional wall motion abnormalities, wall thickness was increased and the RVSP was estimated at 27 mm Hg. The pericardium and the valves were unremarkable; however, the ventricular septum was noted to have dyssynergic motion, again.

DIFFERENTIAL DIAGNOSIS

The patient had signs and symptoms of heart failure upon initial presentation. However, the echocardiographic findings presented a conundrum.

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Figure 1 CT of the pericardium.

Although he seemingly improved with diuresis. The recurrent pleural effusions were exudative in nature, which did not fit with the diagnosis of heart failure. Imaging did not demonstrate any specific diagnosis, and although there was calcification of the pericardium it was not enough to warrant being described as calcified and was thought to fall within a normal range (figure 1).

Figure 2 Ventricular interdependence: classic wave pattern seen in constrictive pericarditis.

Restrictive cardiomyopathy was also considered. Although there are haemodynamic measurements that can differentiate this from constrictive pericarditis,¹ occasionally an endomyocardial biopsy must be performed. Ultimately, pericardial stripping of thick adherent pericardium can solidify the diagnosis of constrictive pericarditis.^{2 3}

TREATMENT

Concern for constrictive pericarditis was raised and a left and right heart catheterisation was performed. The findings were consistent with constrictive pericarditis (figures 2 and 3). Pericardiectomy was performed which revealed a thick, fibrous and densely adherent pericardium constricting the heart (figure 4). Pathology revealed a pericardium of 0.3 cm thickness and chronic fibrosing pericarditis which was negative for malignancy.

OUTCOME AND FOLLOW-UP

His postoperative course was unremarkable and he was discharged from the hospital on postoperative day 5. Subsequent follow-up revealed resolution of the anasarca and slow resolution of the pleural effusions.

DISCUSSION

This case highlights the need for high index of suspicion to rule out constrictive pericarditis, especially in patients without risk factors. In industrialised countries, idiopathic pathology represents the majority of cases, 42–46%.^{4 5} Our over reliance on cardiac imaging to pick up a diagnosis, resulted in unwarranted procedures, that is, VATS. While abnormal septal motion was noted on the first echocardiogram, there was a low index of suspicion for constrictive pericarditis at that time. As this patient did not have a ventricular conduction delay, it was an unexplained finding. Perhaps greater emphasis on this finding with a

Figure 3 Square-root sign with characteristic dip and plateau.

Figure 4 The thick and fibrotic pericardium.

higher index of suspicion, earlier on, may have led to an expedited diagnosis. Also the BNP being slightly above normal has been shown to help differentiate between constrictive pericarditis and restrictive cardiomyopathy. BNP is 'nearly normal' in constrictive pericarditis while in restrictive cardiomyopathy BNP is grossly elevated and averages 825.8 pg/ml.⁶

Recent studies have examined the utility of cardiac MRI.^{7–8} The results from these studies show that this is still a growing field but does not rule in or out a diagnosis. Currently, the latest guidelines from Europe on pericardial disease,⁹ address this modality as yet another tool at a physicians' disposal. However this was unavailable at our institution.

Despite the delay in diagnosis a literature search revealed that this is not an isolated occurrence.^{10–14} Though some studies do outline how to diagnose constrictive pericarditis and restrictive cardiomyopathy via imaging whether with ultrasound,^{15–17} CT¹⁸ or MRI,^{7–8} none of them are 100% sensitive or specific. Demonstration of cardiac pressure waveforms during respiration remains the best way to assess cardiac filling pressures which can lead to a diagnosis.¹⁹ Moreover haemodynamic measurements may not be enough to distinguish between restrictive cardiomyopathy and constrictive pericarditis. Ultimately, tissue may be necessary; an endomyocardial biopsy for restrictive cardiomyopathy or pericardial stripping versus a biopsy for constrictive pericarditis.^{2–3}

The treatment for constrictive pericarditis is the removal of the pericardium. However this surgical procedure carries a very high mortality, currently 6% and not infrequently time is needed for normal cardiac function to return.⁴ Transient constrictive pericarditis has been described, in which the constriction resolved with the medical therapy alone in 8.3 weeks on an average²⁰ and should be considered before sending the patient for a surgery.

In summary, constrictive pericarditis remains difficult to diagnose; however, with high clinical suspicion and newer imaging modalities available, prompt diagnosis is possible.

Competing interests None.

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Learning points

- ▶ A high clinical suspicion is needed to diagnose constrictive pericarditis.
- ▶ A brain natriuretic peptide slightly above normal in a patient with clinical signs and symptoms of florid congestive heart failure should raise the suspicion of constrictive pericarditis.
- ▶ If imaging modalities fail to provide a diagnosis, invasive haemodynamic monitoring may be needed to help exclude the diagnosis of constrictive pericarditis.

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